INTRASPINAL NEOPLASMS IN THE CERVICAL REGION*

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This presentation summarizes the clinical and pathologic features in 179 cases of intraspinal neoplasms in the cervical region in which surgical treatment was utilized. These cases were encountered at the Mayo Clinic from 1914 through 1950. Sections of each of these neoplasms were re-examined microscopically. Excluded from this series of cases were intracranial and thoracic intraspinal tumors that appeared to involve only secondarily the cervical segments of the spinal cord.

PATHOLOGIC FEATURES

The classification of the neoplasms is listed with their incidence in Table 1. The gliomas consisted of 19 astrocytomas, 17 ependymomas, 3 oligodendrogliomas and 2 unclassified gliomas. Of the vascular tumors there were 7 hemangioendotheliomas, 3 hemangiomas, 3 hemangioblastomas and 2 hemangiosarcomas. Among the miscellaneous neoplasms were lipomas, melanomas, lymphomas and sarcomas.

These neoplasms may be classified further according to their location with respect to the spinal canal, the dura mater and the spinal cord itself. Forty-six tumors were intramedullary; although mainly gliomas, they also included 4 vascular tumors, 2 lipomas and a neurilemmoma. Sixty-eight

TABLE 1

<table>
<thead>
<tr>
<th>Classification</th>
<th>Incidence</th>
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<tbody>
<tr>
<td></td>
<td>Number</td>
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<tr>
<td>Neurilemmoma (neurofibroma)</td>
<td>64</td>
</tr>
<tr>
<td>Meningioma</td>
<td>41</td>
</tr>
<tr>
<td>Glioma</td>
<td>41</td>
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<tr>
<td>Vascular tumor</td>
<td>15</td>
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<tr>
<td>Chordoma</td>
<td>4</td>
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<tr>
<td>Miscellaneous</td>
<td>14</td>
</tr>
<tr>
<td>Total</td>
<td>179</td>
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</table>

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tumors were intradural but extramedullary; these consisted of meningiomas, neurilemmomas and vascular tumors. Sixteen neoplasms were extradural yet intraspinal. The 49 remaining tumors could be referred to as "dumbbell tumors." They consisted of 58 neoplasms that were both intradural and extraspinal, together with 11 other neoplasms that were both intradural and extradural. Furthermore, the intraspinal portion of the former group occasionally was both intradural and extradural. Most dumbbell tumors were neurilemmomas. Among others classed as dumbbell tumors were meningiomas, 4 vascular tumors and 2 melanomas.

The segmental level of these tumors is of some interest. The tumor occasionally extended over several segments of the spinal cord, particularly in the case of intramedullary neoplasms. When this occurred, the segment designated as the level was that of maximal involvement. The 4th through the 7th cervical segments were the sites of 55 per cent of all tumors. Of the meningiomas, however, approximately 60 per cent affected the 1st through the 4th segments of the spinal cord. The 5th or 6th segment was the level of 24 of the 64 neurilemmomas.

Syringomyelia was present in 6 of the 10 cases of intramedullary neoplasm in which necropsy was performed. The 6 cases were as follows: 2 of hemangioendothelioma, 1 of hemangioblastoma, 2 of ependymoma and 1 of astrocytoma. Contrarily, in only 1 of the 15 cases of extradural tumor in which necropsy was performed could an associated syringomyelia be demonstrated; in this case, an extensive intraspinal and extradural hemangioendothelioma was present.

**CLINICAL FEATURES**

The entire series consisted of 100 males and 79 females. A girl aged 15 months was the youngest patient, while the oldest was a man aged 76; 8 patients were 13 years of age or younger, yet 4 patients were more than 65 years of age. Of all 179 patients the average age was 38.6. The average age of patients who had meningioma was 42.9, while that of those who had neurilemmoma was 37.6. Patients who had glioma tended to be somewhat younger than the average; indeed, the astrocytomata occurred in patients whose average age was 30.8.

The average duration of symptoms prior to operation for the entire series was 40.9 months. Neurilemmomas produced symptoms 6 months longer, on the average, than meningiomas; the gliomas caused symptoms for an even longer period than the neurilemmomas. Of the common types of tumor the duration of symptoms was longest in the case of ependymomas, namely, 51 months. Symptoms occurred for only several months with hemangiosarcomas, for a somewhat longer period in the case of hemangioblastomas and for a few years when hemangioendotheliomas or hemangiomas were present.

**Pain.** The commonest first symptom was pain; this occurred in 107 of the cases. The initial pain was unilateral, in the midline or even bilateral
and involved the neck, shoulder or upper extremity in 104 cases. Pain appeared in one or both lower extremities as the initial symptom in 3 other patients, and in these it was diffuse, aching and tended to occur during the night. Histories of pain at some time were obtained from all but 26 patients. The pain experienced by the patients in this study usually corresponded in distribution to the region innervated by the nerve roots at the level of the tumor and it characteristically became intensified when the patient coughed, sneezed, strained and flexed his neck. The pain tended to awaken the patient from sound sleep during the early morning and to be relieved by sitting or walking. Occasionally pain was so mild that the patient mentioned it only incidentally or on questioning. Rarely did pain become so severe that morphine was required for relief; it is interesting that this condition arose in a patient who had an intramedullary glioma.

Generally the pain recurred a few times in the same location during the early stages; then as the illness progressed pain usually increased in intensity, became continuous and extended. Facial pain was experienced by 2 patients after the onset of radicular pain. Indeed, for 6 months prior to operation 1 of these patients had facial pain that closely resembled the pain of trigeminal neuralgia, presumably caused by involvement of the descending nucleus or tract of the trigeminal nerve. The character of the pain in this series of cases was essentially the same regardless of the type of tumor. Although the distribution of pain produced by intramedullary tumors tended to be more diffuse than that associated with extramedullary tumors, still radicular pain was experienced by 14 of the 24 patients who had an intramedullary glioma and who noted pain as the initial symptom.

The distinctive pain, therefore, was valuable because it suggested the presence of an intraspinal tumor and was an important aid in the determination of the level of the tumor; however, the character of the pain was related remotely, if at all, to the type of tumor, and use of the presence or absence of pain was worthless in an attempt to predict the type of tumor.

Sensory Disturbances. Disturbances in sensation exclusive of pain occurred in 156 cases. In 28 of these the changes in sensation constituted the first symptom of the illness. The commonest sensory disturbances were numbness, tingling or a diminished perception of one or more sensory modalities. The initial sensory disturbance occurred in one or both upper extremities in 98 cases, while in 50 cases the first sensory disturbance was noted in one or both lower extremities. Sensory disturbances that involved all extremities at some time occurred in 28 cases. Sensory disturbances that always were confined to one or both upper extremities were observed by 38 patients, whereas 35 patients gave histories of sensory disturbances that always were restricted to one or both lower extremities. The occurrence of a sense of constriction about the abdomen or thorax in 14 patients was misleading in determining the level of the lesion. More than 90 per cent of the neurilemmomas, meningiomas and gliomas were associated with objective sensory disturbances.
The distribution of these objective sensory disturbances permitted determination of the approximate level of the tumor in 127 cases. A well-defined sensory level was obtained in 77 cases. It is important to note, however, that in 3 of these cases the sharp sensory level was extremely misleading, for it occurred at the lower portion of the thorax or the upper part of the abdomen. Objective sensory disturbances were deceptively confined to the lower part of the trunk and the lower extremities in several cases of extramedullary tumor, particularly neurilemmoma and meningioma. But intramedullary tumors rarely produced an objective sensory disturbance whose most cephalad extent was significantly caudad to the level of the tumor. All sensory modalities were impaired in 75 cases. The following modalities of sensation were affected in other cases: pain in 61, temperature in 56, vibration in 36, position in 25 and touch in only 12. Sensory dissociation was observed frequently in this series of cases, and it occurred as commonly with extramedullary tumors as with intramedullary tumors.

Weakness. Weakness was experienced by 165 patients, in 27 of whom it was the initial symptom. It was noted first in an upper extremity in about 60 per cent of these cases, while in approximately 40 per cent it occurred initially in a lower extremity. In cases of meningioma, however, weakness occurred initially in a lower extremity more often than in an upper extremity. Weakness was found in all four extremities of 65 patients. Objective weakness was restricted to one or both upper extremities in 36 cases, of which 15 were cases of glioma. Intramedullary tumors produced weakness that, if not limited to the upper extremities, generally was greater there than in the lower extremities; this was observed especially in patients who had ependymomas. It is significant that objective weakness was confined to one or both lower extremities in 24 cases, of which 10 were cases of meningioma. Weakness was present in all but 3 of the 105 patients who had either neurilemmomas or meningiomas.

Weakness of the diaphragm was demonstrated in 11 cases but this sign proved to be unreliable as an aid in the establishment of the exact level of the tumor. Palatal and pharyngeal weakness was observed in 4 cases in which the tumor involved the upper cervical segments of the spinal cord, and in 2 similar cases weakness of the tongue was evident. The following related signs were associated with weakness: muscular atrophy in 76 cases, fasciculation in 36 and spasticity in 33. Weakness had been regarded incorrectly as a residual of acute poliomyelitis in approximately half the children in this series, particularly during the early phases of the illness when weakness was confined to one extremity.

Reflexes. A disturbance in reflexes was observed in 169 cases. In 163 cases this indicated involvement of the pyramidal tracts, and in 49 of these diminished tendon reflexes were also noted in the upper extremities. This combination was of considerable value in a determination of the level of the tumor. In the 6 other cases the biceps, triceps or radial reflex alone was diminished. It is noteworthy that the tendon reflexes in the upper extremities
were diminished in 67 per cent of the cases of intramedullary tumor; in contrast, this occurred in 20 per cent of cases of neurilemmoma and in only 12 per cent of cases of meningioma. The corneal reflex was diminished on one side in 2 cases in which the tumor involved the upper cervical segments of the spinal cord.

Miscellaneous Signs and Symptoms. Impaired voluntary control of the bowel or bladder or both occurred in 107 patients, but this was of diagnostic significance only in that it indicated disease of the nervous system.

Examination of the neck often revealed helpful signs. Stiff neck was observed in 88 cases and its occurrence suggested the cervical region as the site of the tumor, especially when the tumor otherwise gave the erroneous appearance of being situated at a lower level. Tenderness over the cervical part of the spinal column, occurring in 42 cases, was unrelated to the type of tumor, was often diffuse and, if localized, was frequently misrepresentative of the vertebral level of the tumor; still, this tenderness also drew attention to the cervical region as the site of the lesion. A mass in the neck, which at operation was found to be the extraspinal portion of the tumor, was palpable in 14 cases. When associated with other signs of intraspinal disease, this mass suggested the presence of a dumbbell tumor in the cervical region.

Evidence of increased intracranial pressure was observed in 4 cases of glioma, 3 of meningioma and 3 of vascular neoplasm. Definite papilledema could be measured in 6 cases; blurring of the margins of the disk and distention of the retinal veins occurred in 3 other cases and hydrocephalus occurred in a child. Six of these patients gave histories of recent generalized headache; visual blurring or diplopia troubled 4 of the 10, while frequent vomiting was noted by 2 of the patients.

Functional vascular disturbances, particularly cyanosis, coldness and disturbances in sweating occurred in 24 cases, of which more than half were instances of neurilemmoma. These disturbances usually were observed in an upper extremity of patients who had rather advanced disease and commonly were related to disuse. However, specific disturbances in the vascular system appeared occasionally to be caused by the tumor.

Three patients had Raynaud’s phenomenon after the onset of their illness. One of these patients was a meat salesman who had a neurilemmoma that involved the 7th cervical segment of the spinal cord on the right. For 2 years he had observed recurrent episodes of pallor followed by cyanosis of his fifth finger on the right that were induced when he entered a refrigerated room in connection with his work. Intermittent changes in color, which could be classed as Raynaud’s phenomenon, involved the hands of the 2 remaining patients.

Horner’s syndrome occurred in 35 cases, and was observed in 28 per cent of the cases of intramedullary tumor. When associated with extramedullary tumors the syndrome usually was noted on the same side as that of the tumor. Tumors of the lower cervical segments of the spinal cord in this
series of cases did not produce Horner's syndrome more frequently than did those of higher cervical segments.

Nystagmus was horizontal in 24 cases and vertical or rotary as well as horizontal in 6 other cases. The basis of this sign in these cases is not clear. Barbiturates and other drugs had been administered to several of the patients for relief of pain and sleeplessness. Still, when nystagmus occurred, the tumor usually involved the 1st, 2nd or 3rd cervical segment of the spinal cord.

Intraspinal neoplasms in the cervical region may be responsible for symptoms that one perhaps would attribute to extension of the neoplasm into the medulla; however, in this series of cases, such symptoms often were apparently the effect of vascular changes, edema and displacement of the medulla secondary to the intraspinal neoplasm. One patient was distressed by persistent hiccup. Five patients complained of dysphagia and 4 patients stated that they recently had become unable to speak distinctly.

Ten patients gave histories of dyspnea. This apparently resulted from weakness of the respiratory muscles and the disease usually had become advanced by the time this symptom occurred. A history of dyspnea rarely was obtained from the patients examined during recent years.

LUMBAR PUNCTURE WITH MANOMETRIC STUDIES AND EXAMINATION OF THE CEREBROSPINAL FLUID

The dynamics of the circulation of cerebrospinal fluid were tested by jugular compression after lumbar puncture in 121 of the 179 cases. Complete subarachnoid block was evident in 53 cases; partial subarachnoid block was demonstrated in 40 more cases and in 28 cases a normal response to jugular compression occurred.

The content of protein of the cerebrospinal fluid removed by lumbar puncture was determined quantitatively in 97 cases. Values greater than 40 mg. per 100 cc. were obtained in 83 cases; this increase was especially prominent in cases of complete dynamic block, in which values of several hundred or even a few thousand mg. per 100 cc. were occasionally obtained. The content of protein did not exceed 40 mg. per 100 cc. in 14 cases.

The quantity of protein in the cerebrospinal fluid usually exceeded 40 mg. per 100 cc. when dynamic block was absent; conversely, dynamic block often occurred when the protein content was 40 mg. or less per 100 cc. Both abnormalities were present in 65 cases. But 6 other cases were most unusual in that the content of protein of the cerebrospinal fluid did not exceed 40 mg. per 100 cc. nor could any dynamic block be detected.

The cerebrospinal fluid contained fewer than 5 leukocytes per c. mm. in 79 cases, whereas in 31 cases there were 5 or more leukocytes per c. mm. The number of leukocytes rarely exceeded 10 and counts of 20 to 30 were obtained in only 3 cases.

It is noteworthy that in 19 cases lumbar puncture was followed by an increase in intensity of pain, increased weakness or progression of sensory
impairment leading occasionally to the appearance of a definite sensory level.

SUMMARY

The pathologic and clinical features of 179 cases of verified intraspinal neoplasms in the cervical region have been presented. These neoplasms consisted of 64 neurilemmomas, 41 meningiomas, 41 gliomas and 33 miscellaneous tumors. Sixty-eight tumors were intradural, 46 intramedullary and 16 extradural; the remaining 49 lesions could be termed “dumbbell tumors.”

Patients who had meningiomas were usually older than those who had neurilemmomas; the latter were usually older than those who had gliomas. The average preoperative duration of symptoms was 40.9 months. Ependymomas, however, produced symptoms for 51 months on the average.

Sixty per cent of the patients experienced distinctive pain as the initial symptom. Pain was valuable in suggesting the presence of an intraspinal tumor and important in a determination of the level of the tumor; the presence or absence of pain and its character were related remotely, if at all, to the type of tumor.

The reflexes were altered in all but 10 cases. The tendon reflexes in the upper extremities were diminished in 67 per cent of the cases of intramedullary tumor, while 20 per cent of neurilemmomas and only 12 per cent of meningiomas were associated with such changes in reflexes. Weakness and sensory disturbances could be demonstrated in the vast majority of patients, of whom an appreciable number had signs deceptively limited to the lower extremities. Stiff neck, tenderness over the cervical region of the spinal column and a palpable mass in the neck were often present and suggested the cervical region as the site of the neoplasm.

Clinical features have been described that ordinarily are not expected to be associated with intraspinal tumors. Such features appear to be related to the anatomic characteristics of the cervical portion of the spinal cord, including its proximity to the intracranial contents.

Lumbar puncture with manometric studies and examination of the cerebrospinal fluid disclosed either a dynamic block or a value for protein of more than 40 mg. per 100 cc. or both in all but 6 of the cases in which these tests were performed.